May 16th, 1:45 PM

Macroglossia in Inclusion Body Myositis

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Et al.

MACROGLOSSIA IN INCLUSION BODY MYOSITIS

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OBJECTIVE: Discussion of a case of Inclusion body myositis (IBM) associated with macroglossia.

INTRODUCTION: IBM is one of the idiopathic inflammatory myopathies. Exact pathogenesis is unclear but there is an evidence of dysregulation of antigen driven immune response involving T cells. Typical onset is slowly progressive impacting quadriceps often more than hip flexors, ankle dorsiflexors and distal forearm flexor muscles. Swallowing difficulties often are present and mild facial weakness can be seen. Macroglossia has never been reported in association with IBM. In fact inflammatory myopathies of tongue are a rarity.

DESIGN: A case report of a 68 year old woman with 12 year progressive difficulty getting out of chair and inability to twist jar lids. Examination showed forearm flexor and quadriceps atrophy and bilateral face, hip flexion and finger flexion weakness. Clinical diagnosis of IBM was further supported by muscle biopsy findings. One year after the diagnosis, she reported painless progressive enlargement of her tongue resulting in difficulty with chewing. On exam the tongue looked enlarged. MRI of the oropharynx was unremarkable. Tongue muscle biopsy showed non-granulomatous inflammatory infiltrates, myophagocytosis and degenerating regenerating fibers without evidence of amyloidosis. Work up for Sarcoidosis with Chest CT and ACE levels was negative.

RESULTS: Decision to start patient on steroids for idiopathic myositis of the tongue was deferred as our patient is currently enrolled in an IBM clinical trial.

CONCLUSION: Macroglossia in IBM patients has never been reported in literature. Two reported cases of macroglossia were associated with dermatomyositis and granulomatous myositis. It remains unknown whether this is the first reported case of IBM associated with macroglossia or macroglossia due to a second inflammatory myositis.

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